ARCHIVES OF PEDIATRICS

August 1960

Stanford Medical Center, Palo Alto, California

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ALBRO C. GAYLOR, *Publisher*
ROSE E. BODET, *Assoc. Publisher*

JOHN FITCH LANDON, *M.D., Editor*

Vol. 77 No. 8

August 1960

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Published monthly since 1884.
Subscriptions: one year—$7.50; two years—$12.00; three years—$16.00.
Single Copy—$1.00

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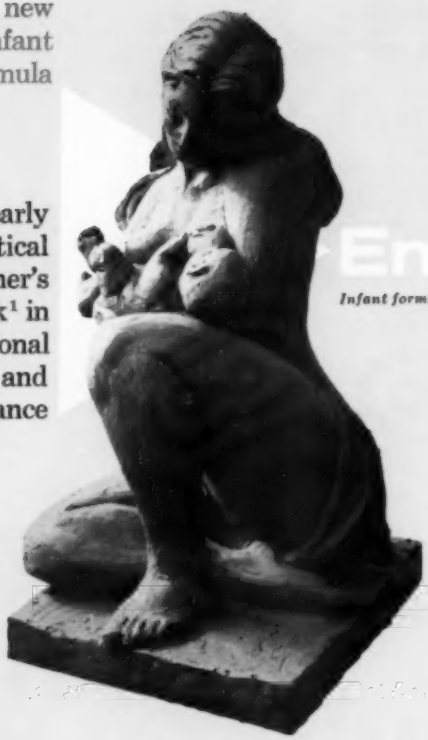

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1. Koessler, H. W.: Arch. Ped. 74:47 (Feb.) 1957.
2. Kahan, H. *et al.*: Arch. Ped. 73:125 (Apr.) 1956.
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The Correction of Clubfoot by Utilizing the Controlled Withdrawal Reflex

MAURICE B. FURLONG, M.D., F.A.A.P.*
GEORGE W. LAWN, M.A., R.P.T.*
New York

ORTHOPEDISTS who treat the congenital deformity known as talipes equinovarus are in general agreement that while it is usually easy to correct this deformity, it is, on the other hand, often difficult to maintain correction. This is true, regardless of which method of correction is used. Hence, treatment over a period of several months, in an effort to prevent recurrence, and observation over a period of years to detect and treat such recurrences promptly, is taken for granted. Even so, a goodly number of infants born with clubfoot have residual interference with foot function so as to require one or several surgical procedures. These, of course, are in addition to those who have residual which interfere with good function but not to such an extent.

Our purpose in presenting this paper is to propose a new and better method of treating the congenital clubfoot. Correction is not only easier and more rapid but we feel much less likely to be followed by recurrence than methods in use today. In fact, although our series is small and our follow-up too brief to be sure, we believe we have practically eliminated this possibility, because our method not only gains rapid correction anatomically but also functionally. The rapid functional correction, not possible with any other method, is gained by our unique approach with which we are able to restore normal muscle balance in approximately two months, in even the most severe clubfoot. This approach involves the use of the withdrawal reflex in a specially modified cast during this period; our technique in carrying out our method is quite simple but most effective.

Before describing our technique, a description of the method of treating clubfoot as it has evolved over the centuries, and especially

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as it has been modified in the past, will give us a better understanding of the many difficulties met in handling this condition, especially as far as maintaining correction is concerned. It will also help us realize why several different techniques based on varying theories have been developed and why there is no unanimity in this area. We will also, before describing our method in detail, explain the theory on which it is based. Any discussion of the etiology or pathological anatomy of this deformity is beyond the scope of this paper; the reader is referred to descriptions by recognized authors in this field of which there are a number.

HISTORY

It is accepted historically that Hippocrates[1] recognized the several components of the clubfoot and all the essential features of the treatment. He said, "Cases of clubfoot are, for the most part, curable, if the deviation is not very great or the children advanced in growth. It is best therefore to treat these cases as early as possible. Most cases are not the result of complete dislocation of the foot in a contracted position." He advised correction "by gentle means" and that the foot be bandaged in a position with an inclination to splay footedness. "Such cases respond more rapidly to treatment than one would think," he said, "but time is necessary for complete success until the part has acquired growth in the proper position". Time has proven the soundness of Hippocrates' concepts. However, he said nothing of the difficulties in maintaining correction and preventing recurrences.

These principles were apparently overlooked until Arcaeus in 1658 described essentially the same method plus a mechanical contrivance to retain the foot in the corrected position. In the 16th century, Ambrose Paré introduced an era of apparatus treatment which ended with Scarpa in the 19th century. We hear little of long term results but must assume they varied from therapist to therapist and patient to patient, depending on the severity of the condition, reaction of the paient, and duration of follow-up treatment, as they do today. In 1803 Scarpa[2] published his Memoir on Congenital Clubfoot of Children. He considered the cause of the deformity to be a dislocation of the forefoot on the head of the astragulus and stated that he had discovered a simple type of apparatus which would gradually restore the normal shape of the foot without inconvenience to the child. He concentrated first on the reduction of the inturned forefoot, then on assisting the peroneals to evert the foot,

and when equilibrium had been restored, on stretching the tendo-achilles to allow the foot to be dorsiflexed. He also emphasized that all soft parts are capable of being considerably elongated without pain or discomfort, when the correcting force is applied in a gradually increasing manner. This of course is the universally accepted sequence of correction of the various components at the present time.

However, despite this philosophy by some, clubfeet were usually treated strenuously and at times radically throughout most of the 19th century, Bruckner[3] used forcible redressment consisting of manipulative maneuvers, carried out in steps and followed by retention devices. Surgery assumed a prominent place in the therapy of this condition in the middle of the century with Stromeyer and Dieffenbach[4] advocating tenotomy of the Achilles tendon and at times cutting the anterior and posterior tibials. Toward the end of this century operations became even more radical; Phelps[5] not only cut the Achilles and tibial tendons but frequently incised the soft tissues, the abductor hallucis, plantar aponeurosis, flexor hallucis longus and short flexors. He finally added to these, osteotomy of the head of the astragulus and a wedge shaped resection of the os calcis!

From this we must infer that results from conservative treatment were not considered very good and more radical measures were believed to be the answer. The surgeons were proud of their successes since they apparently considered correction more in the light of morphological results than function. At times articles would appear such as Barwell's[6] "The Cure of Clubfeet Without Cutting Tendons" in 1863, in which he called attention to his own change in thinking after finding the condition of the feet five years after surgery leaving much to be desired. He found some feet after surgery to be completely valgoid.

Toward the end of the century, therapy gradually became more conservative. In 1889 Bradford[7] stated he believed all except severe neglected cases could be cured by forcible rectification and that a bone operation should be avoided if at all possible. He also believed failure to correct the equinus completely was responsible for relapse more often than anything else. This aspect will be covered in the description of our technique later. In 1895 Walsham and Hughes[8] published their "Deformities of the Human Foot" in which they advocated besides manipulation and retention in casts, tenotomy of the

Achilles tendon in all but the very mild cases. However, Tubby[9] about this time urged conservative treatment with the treatment performed by three stages: (1) correction, (2) retention, and (3) supervision, with particular attention paid to footwear and the manner of walking. He said the ultimate success of the treatment depended more than anything else on the thoroughness with which the third stage of the treatment was carried out.

By the beginning of the century, surgical measures, especially the radical osteotomies, were used less and less frequently. As time went on they were used only in such cases in which the age or degree of deformity precluded conservative measures and it was realized that on the whole functional results by surgery were much inferior to those of conservative treatment. In 1912 Sir Robert Jones stated any congenital clubfoot not associated with a primary condition (such as spina bifeda) should, if treatment is begun in the first week or two of life, be completely corrected by the end of the second month. Jones,[10] like Tubby,[9] emphasized the importance of adequate and prolonged follow-up. In 1928 Adams[11] likewise advocated, besides overcorrection by manipulation and retention for several months, exercises and constant supervision for "some years". Finally, at approximately the same time, almost three decades ago, Kite[12] in this country and Denis-Browne[13] in England, each employing a different technique which he had developed, strongly recommended conservative treatment for the majority of clubfeet.

At the present time, there are three conservative methods used in the treatment of clubfoot. First there is that originated and advocated by Denis-Browne[13] which is familiar to all working in this field and still used by some orthopedists in this country. It has the great advantage of allowing some active motion throughout the entire phase of therapy. While anatomical correction is usually obtainable within a few months it requires close supervision and a long period of treatment—up to one or two years—and the deformity has a strong tendency to recur especially in the adductus and cavus components. As a result, even in experienced hands, this method has to be supplemented by corrective cast treatment. Furthermore, it is estimated from five to eight percent eventually have residual severe enough to justify surgery.

The second method, like the first, originated in England but is not so familiar to workers in this country—it might be termed the

physio-therapeutic method. It consists in applying a corrective lateral splint of aluminum which is removed two or three times weekly at which time passive stretching is done and active motion by stimulation of the peroneals is attempted. This also involves close and repeated supervision and a long period of treatment; the proponents advise special exercises the first few years of walking to develop the proper muscle pattern. Theoretically this method is a sound approach; it permits considerable motion and attempts to develop normal patterns of muscular activity .Its chief disadvantage is the reliance on co-operation from an infant who is too young to give it.

The third method which is used by the vast majority of orthopedists in this country is the use of plaster casts—with or without the wedges originated by Kite. This method is, of course, well known and will not be described in detail. Suffice it to say it involves application of plaster casts weekly or biweekly with gradual correction of the clubfoot and in the accepted proper sequence of abduction, eversion, and dorsiflexion. The method is arduous, requiring many cast applications at frequent intervals for two to six months. Frequently additional series of casts must be applied because of unsatisfactory results. Even then, some have residual interference with foot function which justify surgical attempts to improve this. As in other areas, surgical procedures here, even the relatively minor lengthening of the Achilles tendon, carry a destructive element which must be accepted to obtain possible improvement in function. While this method gives good anatomical correction in the early months, insufficient attention is paid to the problem of obtaining good function and the abnormal muscle balance invariably tends to recur. We believe this accounts for the tight heel cord, the shortened foot and the cavus deformity which are seen so frequently in later years.

Over a hundred years ago Sheldrake[14] said, "when the foot of a young child has been distorted in this manner (clubfoot) it should not be left at liberty after correction until it is able to walk, as the inequality of the muscles of the foot will always give a strong tendency to resume the distorted form." Most authors may be quoted in this same vein today, despite the use of orthodox measures of correction. "The tendency to recurrence is strong," says Turek[15]. "During this period (of several years) any tendency toward recurrence of the deformity should be promptly and thoroughly treated," states Shands[16] in his text book. Kite[17] in a recent article states "Clubfeet are difficult to correct and the clubfoot has a great ten-

dency when corrected to return to its original position. For this reason the correction should be thorough and the foot should be followed for several years, and additional treatment given, should it be needed."

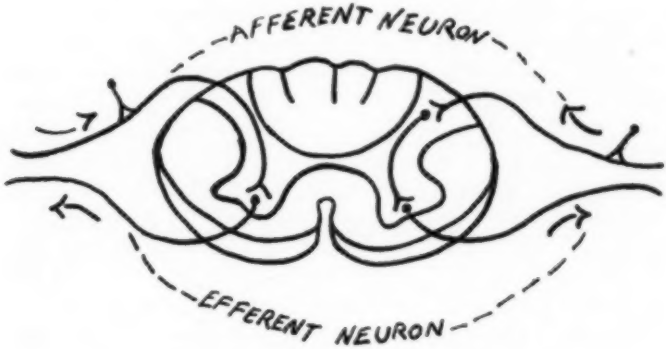
Why is recurrence of the deformity after many months or even years prone to develop? We believe, like many others, that recurrence occurs because the original muscle imbalance is not corrected by treatment. The desirability—nay, the necessity—of restoring the normal muscle balance is being recognized more and more by orthopedists in this field. According to Shands[16] measures should be taken during the second stage of treatment to maintain the correction and to encourage the development of normal muscle balance. He recommends the use of special shoes and further states "walking should be encouraged to strengthen the weakened muscles; such muscle re-education is an extremely important factor in obtaining a permanent cure." Steindler[18] says, "the object of functional rehabilitation is to restore active muscle balance." He mentions message and passive exercises. Holland maintains that correction can be obtained, even though the foot is freed from the splint several times each week, with the patient receiving active and passive exercises on these occasions; in this way the weak, over-stretched muscles can be stimulated and re-educated from the beginning of treatment.

We agree in the necessity of these measures, and furthermore that they should be carried out, as Holland states, from the beginning of therapy and not, as has been the practice, after anatomical correction has been obtained. We should—and can by the use of our unique approach using the infant's normal reflexes—carry out anatomical correction and muscle re-education simultaneously. Indeed, in our method the latter assists us in the former and our primary aim is the restoration of normal muscle balance, the cast or other appliance being merely an adjunct to correction, though obviously a necessary one.

THEORY OF OUR METHOD

The theory underlying our method of correcting clubfoot by reflex stimulation is based on the infant's normal neurological maturation pattern. It is generally agreed that the cerebrum of the newborn, or more accurately the cerebral cortex, exerts no influence upon the segmental apparatus in the newborn. Langworthy states that "the behavior of the newborn is essentially that of a brain stem

preparation". Why this is so can be seen from the anatomical demonstration that the segmental apparatus of the central nervous system, cranial and spinal, is nearly complete at birth, but the chief suprasegmental structures and their projection paths are still unmyelinated and immature. As a result of the lack of cerebral function in the newborn, there is (1) absence of true volitional motor activity and (2) lack of motor inhibition of the segmental apparatus. In other words, the motor reactions at birth are entirely reflex, many of them spinal arcs. One is shown graphically by diagram 1.



DIAG. 1

This is deliberately oversimplified because with the literally innumerable synapses, impulses almost of necessity involve more than one level of the spinal cord. This overflow is illustrated by the reaction of both lower and sometimes the upper extremities on plantar stimulation. However, the reaction does not involve the upper centers in the very young infant.

Specifically, we employ three reflexes in our method of correction of clubfoot. First, and by far the most important, is the flexion or withdrawal reflex which can be demonstrated best in a spinal or decerebrate animal; stimulation of the sole of the foot by pinching, pricking, or applying an electric shock results in a strong contraction of the ankle, knee and hip flexors with simultaneous inhibition of the extensors. This reflex is, of course, not normally present in the older child or adult but is found in the normal newborn with his immature central nervous system. In fact this reflex was demonstrated by Minkowski[19] in the fetus before the third month. It is variable in the early months of life with not only flexion of the ankle

but both flexion and extension of the toes. Toe extension, it is generally agreed, predominates until the second year of life, when it changes normally to a flexion type of response. This type of massive reflex motion in the young infant is ideal for our purpose.

The second reflex we use is the crossed extension reflex; this consists in the contraction of the extensors with reciprocal inhibition of the flexors in the contralateral limb, when plantar stimulation is done. This gives desirable active motion of the extensor groups, and very slight but real joint motion of the opposite limb. In case of bilateral clubbing, with both limbs in casts, this reflex gives us exercise of the extensors groups of the one leg at the very same time we are applying plantar stimulation to obtain the desired action of the flexors of the other leg.

The third is the Frankel reflex; this consists in a re-enforcement of the dorsi-flexion of the foot by resisting the flexion of the thigh as the subject attempts to do so. This reflex is usually used in the adult; in such case it is a voluntary attempt to flex the thigh while deliberately applying a counter force against the knee. In our method, we utilize this by restraining the withdrawal reflex elicited by plantar stimulation. The infant reacts involuntarily; we re-enforce the dorsiflexion of the foot by restraining the knee from flexing on the thigh.

In addition, we are able to avoid by our method what we term conditional anti-corrective reflexes, which we feel operate against correction. By this we mean the resistance offered reflexly by the infant as a result of the repeated cast applications used in the orthodox method to gain correction. Our method requires only one or two cast applications in the first two months by which time we have obtained not only full anatomical correction but have restored normal muscle balance.

DESCRIPTION OF OUR TECHNIQUE

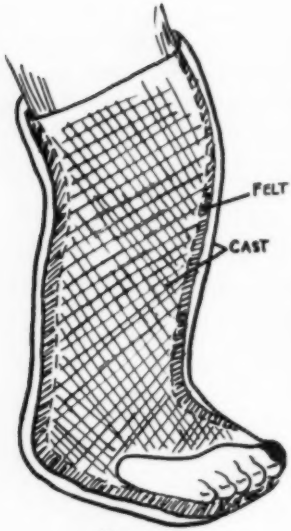
Our method of treating the clubfoot deformity, like that of Tubby and others, consists in three stages: first, correction, secondly, retention; third, observation and supervision. Further we divide the stage of correction into two phases, anatomical and functional, the latter referring to the correction of the muscle imbalance. Chronologically we accomplish full anatomical correction the first month and full functional correction the first two months. To clarify our method of attack, both as to time and to the order of correction of

the various components, we subdivide our anatomical correction into weekly substages. The first week we correct the forefoot, the second the foot, the third the heel varus, and the fourth the equinus. This is not a rigid concept as we accomplish additional foot correction in the third week as we concentrate on heel correction, but it gives a good target date. We consider it important, for example, to accomplish equinus correction by the end of the fourth week before the calf muscles develop much strength; we do this in three, or even two weeks if we can do so.

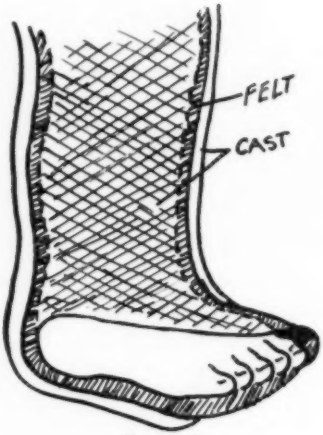
The first stage, that of correction, is the most important, and if carried out properly makes the second and third stages routine. We begin in the usual fashion, with the application of a plaster cast from mid-thigh to the toes, with the foot held in the corrected position, that is, abduction and eversion. This is done after the tight muscles have been stretched manually for five to ten minutes. As in the orthodox method, we obtain as much correction as possible without excessive force. The cast is then converted to an "open" one by removing the portion enclosing the dorsolateral aspect of the foot; this is a most important feature of our method and makes it different from all previously employed techniques. The dorsolateral portion of the cast is removed in two stages. First, the anterior half (back to the cuboid) is cut away; this is done most easily before the cast is dry. Exactly how this is done can be seen from diagram 2. The posterior half, covering the lateral malleolus is removed ten to fourteen days later. This can be seen from diagram 3.

Meanwhile, the day after the application of the cast and the removal of the anterior half of the lateral side of the cast, the first forefoot wedge is inserted; we use felt. Additional wedges are added at intervals of two or three days to take advantage of the great pliability of the foot at this early stage. These wedges, as can be seen from diagram 4, push the forefoot laterally, thereby correcting the forefoot varus and to some extent the forefoot supination. In addition the "open cast" allows some foot motion which we obtain by reflex stimulation—the second and the most important feature of our method which distinguishes it from any previous technique. This feature and how it is carried out will be described below.

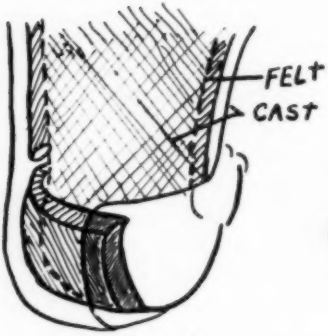
Seven to ten days after the application of the cast, it is cut farther back on the dorsolateral aspect back to the heel, so the lateral malleolus is exposed; meanwhile considerable correction of the foot, especially of the forefoot, has been obtained. Large felt wedges are



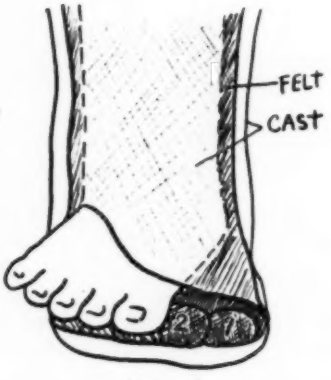
DIAG. 2



DIAG. 3



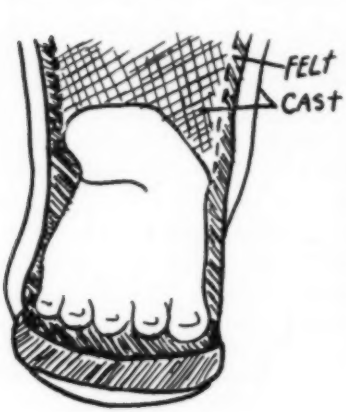
DIAG. 5



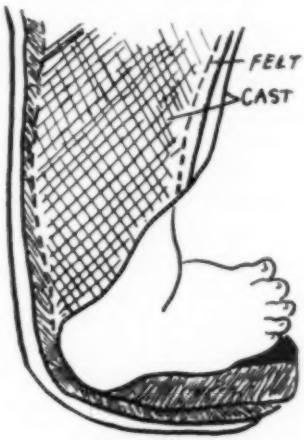
DIAG. 4

then inserted between the cast and medial border of the foot. This can also be seen from diagram 4. The removal of the lateral section of the cast (foot portion) allows the entire foot to swing laterally, thereby correcting the adduction and inversion of the foot. In addition we help correct the supination of the foot by insertion of a wedge under the cuboid bone. The latter tends to prevent the tendency of the foot to return to its former position of adduction and

supination. More correction of the varus heel may be obtained by inserting a felt wedge between the cast and the medial aspect of the heel. Again the deficiency in the cast laterally allows a lateral correction, this time of the heel. This can be seen from diagram 5.



DIAG. 6



DIAG. 7

By this time—the end of the second or third week of treatment—the foot is almost completely corrected anatomically. The procedure thus far has been directed to the adduction, supination, and inversion components and efforts can now be made to correct the plantar flexion component of the clubfoot deformity. This is earlier than in the orthodox method but we believe it is very important to accomplish this before the calf muscles develop their great strength, and is one reason for the success of our method. We do this by further trimming the cast—this time removing the part covering the anterior aspect of the ankle and the lower foreleg. This allows free dorsiflexion as can be seen from diagrams 6 and 7.

With this modification of the cast we can insert a thick plantar wedge between the sole of the foot and the cast. We usually use plaster for this, as it must be quite thick and it can be molded exactly to the desired contour. This, of course, corrects the plantar flexion deformity. In the true clubfoot, dorsiflexion at the onset (birth) is impossible. Attempts to focibly dorsiflex the ankle are destructive to the proper alignment of the tarsal bones, especially the navicular. However, at the point we have now reached in our treatment, the

inversion and adduction of the foot have been corrected and the tarsal bones are in normal alignment. Dorsiflexion of the foot now can be done without fear of structural damage. Normal passive dorsification, however, is still impossible without considerable stretching of the Achilles and resultant pain. Therefore we do not wedge to the limit of passive dorsiflexion. In addition this allows some active dorsiflexion on plantar stimulation; this is an important point in our cultivation of muscle function and balance. Usually there is time to insert a second wedge—here we use thick felt—before the infant outgrows the original cast. During this entire period, of course, reflex stimulation as well as passive stretching is carried out by the mother.

At this point we will describe in detail the unique way we employ reflex stimulation to help correct the deformity and, more important, to develop normal patterns of motion. The latter, we believe, is the crucial factor in a permanent cure. Beginning the day after the application of the first cast, we begin stimulation of the withdrawal reflex by stroking the foot—usually the plantar surface—at frequent intervals during the day. We use a bare applicator, inasmuch as the point cannot be too blunt. The cast, being closed dorsally and medially but open laterally, prevents the foot from inverting or supinating and forces it to move in the desired direction of eversion, pronation, and abduction, and, later, dorsiflexion. Over a period of weeks this causes the foot motion we desire, which imitates the pattern of the normal infant, born as he is with a straight and very flexible foot. Although reflex stimulation is very simple, it is such a vital part of our method that we instruct and check the nurse and later the mother in its execution, since they obviously must be responsible for its being carried out. We have the nurse or mother do this ten or twelve times on perhaps twenty or twenty-five occasions during the day. Usually we tell the mother to carry out reflex stimulation before and after each diaper change. This is a convenient time and way for her to remember. We employ the Frankel reflex in conjunction with the withdrawal but not until we obtain some ankle motion on plantar stimulation. This is usually after three or four weeks of treatment at which time good passive dorsiflexion of the foot is easily obtained and we can expect some active dorsiflexion on reflex stimulation.

In addition, we have the mother do passive stretching in the direction of correction. She does this about six times—four times a day; the exact direction depends on the stage of correction and the foot

contour. In the beginning—the first two weeks—she stretches the tibials as well as the intrinsic foot muscles by abducting the foot, mostly the forefoot; this is done gently. After the correction of the adduction and supination defects, she dorsiflexes the foot on the leg, again gently. This stretches the Achilles tendon and, to a slight extent, other muscles and tendons. We emphasize gentle stretching feeling that vigorous stretching in the past has contributed to the incidence of rocker foot and pes planus.

By the time two or three medial wedges have been inserted between the foot and cast and the thick plantar wedges are also inserted, a month has elapsed. In some instances, removel of the thick felt padding will allow the use of the same cast another two or three weeks, long enough to gain full functional correction. In most cases, it is easier to apply a second corrective cast, especially if more correction of the heel and foot are desired, as with a severe clubbing. The same principles of correcting by wedging while allowing some active motion should be followed. The infant then enters the second stage of treatment, that of retention. During this stage our chief aim is to retain not only full anatomical correction but also the restored normal muscle balance. Our main efforts in maintaining the latter are directed to preserving the balance between the foot flexors and extensors. The original imbalance between the tibials and peroneals we feel will not recur in any event after we have reached this point.

Originally on plantar stimulation the motion of the foot is very slight; in fact, it is practically limited to the toes, due to the marked tightness of the muscles and tendons, medially, and the overstreching of their antagonists. However, allowed and encouraged in its lateral excursion by the "open" cast, the foot gradually increases its ability to move in this direction, so that, by the end of the first month of treatment, the foot has developed a normal pattern of motion, although the range is not as great as in the normal foot.

We should mention here a few points in technique we have found very helpful. First, we use heavy felt (¼") instead of stockinet to encase the thigh, leg, and also the foot, with the exception of the medial aspect of the heel and the foot. We feel the felt is resilient enough to allow a little motion and is preferable to the adherent skin-tight cast which is designed to truly immobilize. In addition, this makes pressure points less apt to occur and also allows us to use the same cast an extra two or three weeks. By removing the

felt padding, the cast is enlarged and can be used longer, despite the rapid growth at this time. Second, we use an additional triangular-shaped piece of felt over the anterior ankle so that, on its removal after four or five days, we can get better motion of the foot on reflex stimulation as well as greater range of passive motion. Thirdly, we bivalve and remove the cast after seven to ten days to check for areas of pressure; it is very easy, of course, to replace the cast on the extremity and fasten with adhesive. This allows us to inspect and adjust the cast as desired on this, as well as subsequent occasions, with less stress and strain on both operator and infant. In addition, should edema develop, as is practically unavoidable at times, removal of the cast for twenty to thirty minutes allows it to subside. We have not found it necessary to remove the cast at other times except at regular office visits. Fourth, we sometimes find it helpful to use part-time wedges. In the first two weeks when the tendency of the foot to supinate is very strong, the lateral displacement of the foot by the medial wedges may allow it to escape from the confinement of the medial section of the foot portion of the cast and resume its marked supination. This can be obviated, if necessary, by removing one or two of the medial forefoot wedges during the day so the medial forefoot is again restrained by the cast. These can be inserted at night to obtain the further stretching desired. Lastly, we use a plantar wedge part time as routine, to obtain greater stretching of the Achilles tendon without discomfort to the infant. When we have gained good anatomical correction of the adduction, supination and inversion—usually in three weeks—we add, as described above, a thick plantar wedge of plaster. We then, beginning immediately, have the mother insert a thick felt wedge each night to stretch the Achilles tendon further and thereby very soon gain normal dorsiflexion on passive motion. The removal of the felt wedge allows more foot motion during the day when reflex stimulation is carried out.

During the second phase of treatment, namely, retention, we continue the use of a splint to maintain anatomical correction—or slight over-correction—just as the orthodontists use retainers for teeth which have been straightened. The teeth retainers are originally worn full time, then twelve hours out of twenty-four, etc., so that the teeth "set" in their new location. We continue the leg splints for approximately four months for this purpose, usually for one month full time, and then three months part time, at night and during naps. As in the first phase, we have the mother continue to use

reflex stimulation both while the foot is splinted and later during the period the splint is removed. Our theory is that in this way the correct muscle pattern and balance, originally merely spinal, becomes impressed on the suprasegmental apparatus as it develops. In addition, we have the mother continue to use gentle passive motion three or four times daily to maintain flexibility of the foot and ankle joints.

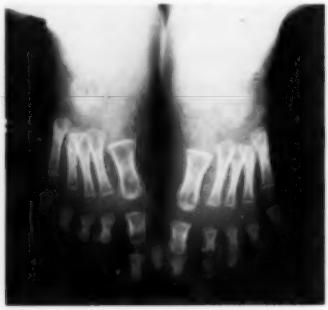
Our only difficulty in retaining full correction once gained has been in maintaining the normal range of dorsiflexion of the foot. We have corrected the adduction, inversion and supination of the foot and have retained correction of these features and maintained good reflex patterns with discontinuation of all treatments—casts, stretching exercises, etc.—after two or two and a half months. However, the infant loses the excellent dorsiflexion of his foot gradually over the next few months—unless splinted—as the Achilles tendon tightens to resume its former shortened character. Apparently this is because the dorsiflexors of the foot, even with the strengthened peroneals, are not able to maintain good foot dorsiflexion actively and the stronger foot extensors tip the balance in the direction of plantar flexion. Theoretically, passive stretching should be sufficient to keep the Achilles tendon lengthened, and no treatment should be needed after two months. In practice, however, the infant's natural tendency towards plantar flexion, exaggerated by the originally short Achilles tendon and the infant's invariable reaction to pain, results in the very thing we are trying to avoid. He pushes against the exerted force, and the stretch procedure becomes, in its effect, a resistance exercise procedure, strengthening and shortening the gastroc-soleus muscles. We therefore use the retention splint for approximately three months until normal dorsiflexion becomes natural to the infant.

We agree with the importance and the necessity of observation and supervision in the third phase of management of the deformity at the present time, although we feel that our method produces a real and permanent cure. We believe that in the future after our method proves itself, recurrence will be very unlikely and will not have to be feared. We therefore concur, at the present time, with the recommendation of Steindler—a period of observation over a number of years. We have not found any indication for the use of special shoes or any special exercises, including guiding the child in walking in a certain pattern.

DISCUSSION

Our results in treating five patients by our method has given excellent results in every one except the first (patient No. 1) in whom our use of reflex stimulation was not begun until the infant was over five months old. This patient was treated in orthodox fashion for the first few months. He has a good result; he has some restriction of dorsiflexion of the feet at the ankle due to short Achilles tendons but has normal reflex patterns and has shown no tendency to relapse at the age of thirty months.

Patient No. 1
C.T. X-ray, age 1 day



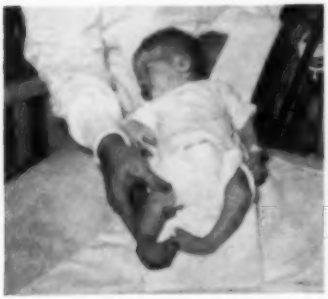
Patient No. 1
Bilateral clubfeet
X-ray, age 18 months

The remaining patients, ranging from eight to twenty-eight months, were treated by our method using reflex stimulation from birth and have excellent results, both cosmetically and functionally. The results in the two infants with a unilateral deformity are such that it is difficult to tell which was the affected foot. They have normal development of the foreleg muscles so the affected leg is equal in size to the one normal from birth; the feet are of equal size including length. All have retained good reflex patterns of foot motion, as well as normal flexibility.

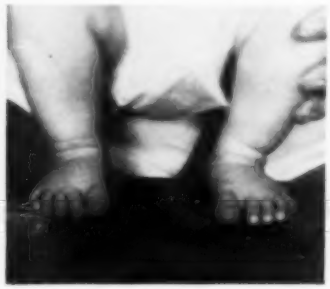
Every good article, after presenting the aims of the present, advances speculation as to the future possibilities in the area under discussion. Our first would be in the use of our method in older children, especially those who had not been treated. Older children with no previous treatment have been reported treated with good results by Kite, using orthodox casting method. In the patient mentioned above in whom we began to employ our method late—that is, at five months of age—we did succeed in correcting the abnormal

reflex patterns and have them remain corrected. Thus we feel it is possible, in fact, probable, that our method might be used successfully in not only the newborn but in older infants and children.

We might speculate further on the use of our method of reflex stimulation—or some modification—in conditions other than the clubfoot deformity. For example, we feel use of our technique might be helpful in some infants and small children with weakness of the lower extremities from poliomyelitis. Happily, poliomyelitis is not seen as frequently as formerly but in instances of weakness of the lower extremity stimulation of the foot, while it is restrained by a mechanical device from assuming an abnormal position, could well facilitate recovery. We refer here to a patient too young to cooperate in physiotherapy. We believe it would in such instances reduce the residuals so commonly seen of marked muscle imbalance—so difficult to correct after poor patterns of motion have developed.



Patient No. 4 C.C.
Photo, age 2 days



Patient No. 4 C.C.
Photo, age 7 months

Cerebral palsy is another condition in which our method might be of help in assisting the infant to develop a good muscle pattern in the lower extremities. The ideal patient in this category would be the congenital spastic diplegic on his way to the development of deformed lower extremities. In theory, the use of our method to encourage good muscle balance in the affected extremities, using splints or casts, would be effective in this condition as well as in the clubfoot deformity.

CONCLUSION

We have presented an original and dynamic method of treating clubfoot, a method which corrects the deformity anatomically and

functionally at the same time. We have presented the results in our small series. We have advanced the possibility that employment of the same principles used in our method might be helpful in conditions other than the clubfoot deformity. Perhaps our method will open a new avenue of approach in the management of these conditions in the earliest months of life. Perhaps the immaturity of the neurological apparatus of the infant can be an advantage in treatment instead of a hindrance as it now is. Perhaps experimentation will show that a new method based on the principles we suggest will be effective in the early months of life and will therefore allow us to use our current methods to better advantage in the latter years of infancy and childhood. Such is our belief and our hope.

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Amblyopia Detection in the Practice of Pediatrics

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To most pediatricians, eye problems in children belong strictly in the domain of their colleagues in ophthalmology, and they are usually quick in referring these young patients, if they suffer from more than a simple conjunctivitis. In general this may be the right attitude. However, there is one eye condition, amblyopia ex anopsia, that the pediatrician should be acutely aware of, because he is in the fortunate position to detect it in the early stages when treatment is effective. He only has to think of it, and routinely test his patients' vision as early and as reliably as possible. The earlier amblyopia is detected, the greater are the chances of successful treatment. The possibilities of cure are considered very good before the age of 5, fair between 5 and 8, doubtful between 8 and 10, and almost nil after 10.

Amblyopia ex anopsia, or "lazy eye", is a very common condition. Its incidence is given as about 2% in the literature. It often is an inherited trait, and may be seen in successive generations. Although its nature is not completely understood, we know that we are dealing with an eye that is structurally normal, but has a deficiency in form sense. Amblyopia may be due to suppression or underdevelopment of this important retinal function in cases where there is interference with normal binocular vision in early childhood. This interference may be a strabismus, where the image in the deviating eye does not fall on the macula. Or, it may be an anisometropia, where the eye with the higher refractive error receives a less distinct image, and, therefore, does not learn to see clearly. In some rare instances, no obvious cause for the amblyopia can be found.

In our highly developed industrial civilization, many occupations require perfect vision in both eyes, and we really cannot afford to neglect this preventable form of visual deficiency. A patient with amblyopia of any significant degree lacks not only fine depth perception, but often is subject to eye strain when excessive demand is made on his good eye which carries all of the burden. He may

also become seriously handicapped if the good eye should be lost in later life because vision in the amblyopic eye usually cannot be improved.

In recent years some advances have been made in the treatment of amblyopia by intensive pleoptic training. While cases with central macula fixation of the amblyopic eye usually respond well to simple patching of the good eye and correction of the refractice error, amblyopes with extramacular fixation may be made worse by this procedure, and should be given active stimulative "waking up" exercises of the macular functions by means of special instruments. Under direct observation of the fundus a circular area around the macula is illuminated with a strong, dazzling light, leaving the macula itself unexposed. This will produce a temporary "ring scotoma" covering the false macula, and leaving the real macula receptive for direct stimulative training. Good results, in otherwise hopeless cases, have been reported with this method.

In general, treatment does not present too great a problem if we can start it early. How can we detect amblyopia early enough? If the child has a strabismus, the possibility of a lazy eye is obvious, and should always be investigated. If the eyes are straight, however, the only way to detect amblyopia is by testing the visual acuity. To do this, the child has to reach a certain maturity. Before the age of 3½ we can not really test for visual acuity, but only observe the behavior reaction of the child to the object with which he is confronted. Any reaction, when it occurs, may be considered as proof that the object has been visualized. The grasp reflex, which is the most common type of reaction, matures so much earlier in life than the vision, that it may very well be used to test the latter. In assessing the vision in a small child, each eye is tested separately, using a patch to cover one eye, and the reaction (grasp reflex) is compared. A definite difference in behavior will be due to a difference in vision in the two eyes, and will, therefore, be of the greatest significance.

To estimate vision in children below 3½, we have been using the *Candy Bead Test*. This test and screening procedures have been fully described in an article in "Modern Problems in Ophthalmology", Volume 1, and Supplementa Ad Ophthalmologica, Fasc. 47, Page 405, 1956. Cake decorettes are used as objects which the child picks up with one eye patched. The reactions with either eye opened are compared. This "Home Eye Tests For Pre-School Chil-

dren" Kit consists of an E Chart*** with the description of the test on the back, a patch, and a gelatin capsule with cake decorettes. It can be handed out to interested mothers by the pediatrician and general practitioner, or by the nurses in well-baby clinics and nursery schools, etc. It has been our experience in distributing over 4000 of these kits through these channels, that only a relatively small number of mothers will do this test and report back to us unless the doctor or nurse takes a few moments to explain to them the importance of this examination. There is no doubt that the test does pick up amblyopia in the earliest possible stage, although such an amblyopia must be of a rather significant degree (more than 20/70). It is precisely these amblyopias with which we are concerned at this particular age, and, therefore, the test should be used as widely as possible.

After 3½ visual acuity can be tested in most children by the illiterate E Chart. This should really be done by all pediatricians, and not left to the responsibility of the mothers. A routine screening with the E Chart in nursery schools should also be encouraged, and, at present, is being tried in several areas. The most important step in amblyopia detection is vision screening in kindergarten. If it were done reliably in all schools, the problem of amblyopia would most likely become insignificant within a few years. In vision screening from first grade up, the emphasis gradually shifts from amblyopia detection to visual performance in relation to the demand of the school work; in other words, detection of the refractive errors and muscular imbalance. These latter conditions should, of course, be investigated and corrected because they will help the "learning child". But there is no doubt in my mind that early screening for amblyopia is much more important, because we are searching for a condition that cannot be corrected in later life.

One important practical point in visual testing has to be emphasized, particularly when we are on the lookout for amblyopia in the young child. Children who do not see well with one eye, usually do not realize this defect, and have adapted to it so well, that casual observation will not detect it. They have learned so well to use the good eye in any situation that they have become very clever at peeking around any occlusor device, unless they are very closely watched. In the usual school screening—using cups or cardboard occlusors—it happens, therefore, time and again, that an amblyopia is missed, often for several years, until it is too late for effective treatment. A new occlusor** has recently been devised (first described in the American Journal of Ophthalmology, Volume

48:847, December, 1959) to make screening foolproof (Fig. 1). It not only eliminates peeking, but also makes screening more rapid. The mask is molded of white plastic and has two 8 mm. vision holes. This size allows for the variation in the pupillary distance and prevents the stenopeic effect—which is without influence in amblyopia anyway. A dark brown moustache-like movable flipper arm that allows only one vision hole open at a time is mounted as shown.

Fig. 1. Simple occlusor mask.

When one hole is open, the other one is securely shut. The oblique position of the flipper arm allows the examiner standing at the vision chart to tell from afar which eye is being tested. This eliminates a helper who ordinarily stands next to the child and covers the eye. The child can move the flipper arm himself, or another child can do it for him. The mask is very well accepted by children of all age groups who think it is great fun, particularly when the screening is done around Halloween time. The standard mask fits any child from 5 to 12, but they can be ordered to fit smaller or larger children. The elasticity of the temples holds the mask well in place and makes it easy to slip on and off. In removing it, the mask should be pulled downward towards the chin to avoid touching the eyelids with the ear pieces of the mask. The masks can be scrubbed with soap and water, and therefore, are very easily cleaned (much easier than the trial frames we use for refraction in the office). We have never encountered any transmission of disease in our screening with this mask. Recently a modification of the flipper has been made that also allows testing for excessive hyper-

opia (plus sphere test) (Fig. 2). Two plus 2.0s lenses are inserted in the flipper in such a way that they cover both vision holes, when the flipper is placed horizontally. Only hyperopes of more than plus 2.0 will be able to read the chart through them. In the oblique position the flipper is used for testing each eye by itself in the same manner as with the simple type. For examiners who routinely do their plus spheres test, this type of flipper is a great time saver.

Fig. 2. Flipper with + sphere lenses used in testing for excessive hyperopia.

Our school nurses* have screened over 10,000 grade school children with this device during the last two years, and picked up 25% more amblyopias than with the conventional occlusor devices that were used on previous testing.

SUMMARY

Amblyopia ex anopsia, a common preventable form of visual deficiency which can only be treated effectively in early life, is discussed. The pediatrician should be very conscious of this condition, and help in its early detection.

A vision testing kit for preschool children is mentioned, and a new occlusor mask is described which makes screening foolproof and rapid. 25% more amblyopias were picked up with this device than with conventional means of occlusion.

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* We are grateful to Mrs. Mildred Thompson, Supervisor of Nurses of the Palo Alto Unified School District, and Mrs. Margaret Clark of the Sunnyvale School District, and their staffff of school nurses, for their excellent cooperation. They not only supervised the actual vision screening and compiled the statistics, they gave us the necessary encouragement to go ahead with what appeared to be a worthwhile sight-saving project. We also wish to thank Dr. Harry Jennison, pediatrician at the Palo Alto Clinic, for his interest and help with this manuscript.

** The masks may be ordered by writing to Occlusor Masks, P. O. Box 199, Station A, Palo Alto, California. Patent pending.

*** The E Chart can be obtained from Drexel Printers, 546 College Avenue, Palo Alto, California. We used Duke Laboratories' elastoplast eye occlusors and filled a gelatin capsule with some cake decorettes obtained in the grocery store.

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Further Studies in Complicated Cerebral Palsy

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COMPLICATED cerebral birth palsy is defined as nonfamilial paralysis or paresis with epilepsy or mental deficiency manifesting itself early in life, usually toward the end of the first year after birth.

This paper is being written with the express purpose of bringing before the medical profession a new concept of the pathology of complicated cerebral birth palsy. In sixty-four children who have had craniotomy performed because of this condition, I[1,2,3] found cerebrbal venous angioma at the lower angle of the Sylvian fissure on one side in sixty-one children. One boy with hydrocephalus had a subarachnoid cyst in the region of the cisterna magna, one child had a subarachnoid cyst and porencephaly in the temporal region, one child had bilateral subarachnoid cysts and one child had microcephaly.

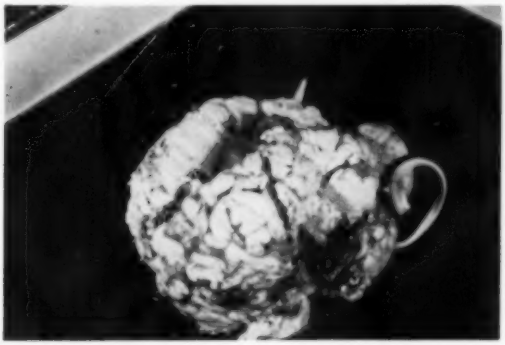
In 1843, Little of London stated that difficulty of labor may be a causative factor.[4] However, in 1862 he published a second monograph on the subject, stating that, as a result of further investigation of a large series of postmortem examinations, he was of the opinion that cerebral spastic paralysis was due to intracranial hemorrhage at the time of birth in almost 75 per cent of the cases.[5] In 1885, Sarah McNutt confirmed this opinion by careful study of pathologic specimens.[6] Beneke,[7] Pott,[8] Mayer,[9] Holland,[10] Schaefer,[11] Schwartz,[12] Saenger,[13] Capon,[14] Barnett,[15] and others have have contributed valuable postmortem observations. Fischer,[16] of Basel, wrote that his postmortem observations at the Institute have convinced him that "the 10% of deaths during the first month of life are chiefly due to cerebral birth injuries". Huenekens,[17] in an article published in 1924, maintained that "the recognition of cerebral hemorrhage of newborn is a most neglected phase in their care and yet is a most important one".

The monumental work of William Sharpe[18] on cerebral spastic palsy should be brought to the attention of the medical profession.

* Director of Neurology and Neurological Surgery, Parkchester General Hospital.

What follows is, in great part, adapted from his writing on the subject.

William Sharpe, over a period of fifteen years, investigated a large number of cases of acute extensive intracranial hemorrhage in the newborn and a large group of children with varying degrees of spastic paralysis, epilepsy and mental retardation. The hemorrhages in the newborn were confirmed by lumbar puncture, cranial operation and necropsy. In selected cases of the chronic type the diagnosis of former intracranial hemorrhage, most probably occurring at the time of birth, was confirmed by operative and postmortem observations.



Massive Angioma in a case of Complicated Cerebral Palsy.

Dr. Sharp was so impressed by these data that he advised a lumbar puncture in suspicious cases within twelve to twenty-four hours after birth. This was tried on a first series of 100 consecutive newborn babies as an accurate means of diagnosing the acute condition.

Intracranial hemorrhage of the newborn is of three types:

(1) Massive hemorrhage, which manifests itself by stupor, respiratory difficulties, refusal or inability to feed, muscular twitching or convulsions. Without operative intervention, the children die.

(2) Mild intracranial hemorrhage—i.e., a degree of loss that can be entirely absorbed. The acute signs, if any, are temporary, and the child makes an excellent recovery. If anything abnormal is noted during the period of absorption, it is usually soon forgotten.

(3) Hemorrhage involving loss of an insufficient quantity of blood to cause death, yet too much blood to be absorbed entirely. Babies with this type are the ones who only apparently recover

within two to four weeks. They are sometimes considered normal for a number of months. Within a year, however, it is noted that the child is not developing physically at the normal rate. It is retarded in sitting, in holding the head up and later in walking and talking. Within the last few months of the first year a spastic condition of one or more limbs is noted. Diplegia, hemiplegia or paraplegia, with or without mental defect, is later noted.

William Sharpe's observations on lumbar puncture in 500 consecutive cases were as follows:

(1) In 45 babies, or 9 per cent, there was bloody or bloodtinged spinal fluid. Of these, 33 were boys, 22 were first born children and only 4 were premature.

(2) The hemorrhage is usually over one side of the brain; therefore, the affected function is usually more marked on one side.

(3) In cases of mild involvement the clot may be absorbed completely, with little or no resulting pathologic change. However, epilepsy may develop later in such cases, or other signs of cortical irritability or instability may appear.

(4) Intracranial hemorrhage not related to the pyramidal tracts may affect the sensory or mental function of the patient.

CONCLUSIONS

The concept of a cerebral cause for cerebral palsy is self-evident. However, the neurosurgeons, except for Wm. Sharpe, have done little or nothing, to help these unfortunate children or their parents. This does not take account of the work that is being done on adults.

In this series of cases, sixty-four children have undergone craniotomy. The sexes were about equal. The youngest was 2 months and the oldest was about 13 years of age. Two children had a bilateral craniotomy.

Clinical Signs and Symptoms that were exhibited in these cases were cerebral paralysis usually of all four limbs of the spastic or athetotic type, mental deficiency in almost every case, inability to hold head up or sit up, strabismus in one, or both eyes, amblyopia, drooling of saliva in about half the cases, difficulty in swallowing, difficulty in respiration with stertorous breathing, and epilepsy. The epilepsy was of pyknotic variety, petit mal, focal or generalized type.

Causes of Cerebral Palsy in this series of cases: Birth injury by precipitate delivery, prolonged labor, instrumental delivery, haemorrhagic disease of the newborn, blood dyscrasia and possible encephalitis. These all resulted in formation of some clot at the lower angle of the Sylvian fissure with subarachnoid adhesions. This venous stasis caused angioma and hypoxia of the brain.

Basis for Operation: The child must have physical disability with little or no progress with medication or rehabilitation. Mental disability and epilepsy should be no deterrent to operation. In fact, the epileptic picture may often give an indication as to localization. If the child shows that the spasticity is more marked on one side, and most of them do, then mental defect should not stand in the way of operation. Increased disability on one side usually indicates one sided cerebral pathology.

Skull x-ray will often give an indication as to which side should be exposed at operation. The venous channels will be more marked on one side and, occasionally, one will find a venous sinus in the region of the pterion on one side. After all the evidence is weighed, then the side for craniotomy is chosen. Sometimes, only minimal evidence is available for aid in making this decision; in such a case the left side should be done in children who come from a right handed family. Often, there is no indication of "handedness" in these children.

Craniotomy: A linear incision is made three inches long, centered over the pterion and running in the coronal plane. The skull often is definitely of a bluish cast. The skull is opened to the size of a silver dollar. The dura may be normal or may also seem to have a bluish discoloration. Occasionally, the middle meningeal veins are enlarged and tortuous. These vessels are cauterized. The dura is opened in a stellate manner. However, one should be very careful in lifting the dura for the arachnoiditic adhesions may tear into a venous sinus or enlarged vein. The angioma is found in relation to the lower angle of the Sylvian Fissure. It may be one of three varieties:

(a) A venous sinus which may take any shape and occasionally is circular.

(b) A venous lake with tortuous enlarged vessels feeding into it and small arteries running over the surface of it.

(c) One or more enlarged veins feeding into a central vein. The brain is of a reddish-blue color and is at first non-pulsating. There

are arachnoiditic adhesions in relations to the angioma and very little flow of cerebro-spinal fluid. The brain may begin to pulsate immediately after the dura is opened and sometimes pulsates while the dura is being opened.

After the angiomatous vessels are cauterized with electrocautery, the brain will become pinkish-red within a number of minutes. After hemostasis, the temporal muscle is closed but dura mater and temporal fascia are not closed. Skin is repaired by separate silk sutures.

Mortality: Five children out of sixty-four cases have died a few days post-operatively. Cerebral oedema was tne cause of death in one and broncho-pneumonia in the others.

Complications: Two children tore open their incisions at home and brain herniation resulted. One of these ihildren subsequently died of brain abscess 1½ years after surgery.

SUMMARY

Sixty-one cases of venous angioma in the region of the pterion were found in sixty-four cases of complicated cerebral palsy which came to operation.

In the children who survived operation, all improved. Some of the children improved slowly and only in small things such as muscle control, muscle power, diminished spasticity. All who had difficulty in swallowing, and breathing overcame these difficulties. All who had amblyopia showed some improvement. Spasticity was always diminished and so was athetotis. Epilepsy never returned in the majority of children and was definitely diminished in others. Strabismus was improved or completely gone after operation. Drooling was diminished in most cases. Mental status was improved so that in some cases the children began to say words, became playful and showed signs of active intelligence. Some children are beginning to walk who could not sit up by themselves before operation. All have shown some progress and some have shown remarkable progress. Rehabilitation has an important place in the post-operative care of these cases for the operation only makes these children more amenable to rehabilitation. Marked improvement should be possible in a majority of cases over a prolonged period. Glandular extracts of Pituitary and Thyroid and Vitamin B1, B6 and B12 play an important part in post-operative care. It is my opinion that the pathology is a definite entity and

that it indicates a means of attack which has helped some of these children. The procedure is deserving of further trial.

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1882 *Grand Concourse, New York 57, N. Y.*

The Dental Management of the Patient With Sickle Cell Anemia

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SICKLE cell anemia is a chronic hemolytic anemia characterized by sickle-shaped red blood cells and occurring almost exclusively in the negro race. The condition is due to an inherited abnormality of hemoglobin transmitted as a dominant characteristic. Homozygous sickle cell patients comprise 0.2% to 0.5% of the negro population in the United States and heterozygous sickle cell patients comprise 5.0% to 10.0% of the negro population in the United States.[1,2]

Sickle cell hemoglobin has the property of forming crescent-shaped cells in the reduced state. The anemia associated with the condition is the result of red blood cell destruction. The other clinical manifestations of the condition are thought to be due to thrombosis in the arterioles and capillaries as a result of the impaction of the pathologic cells.

I. Signs and symptoms are: A. Anemia—moderate to severe. B. Episodes of abdominal pain which may simulate other conditions. Vomiting also occurs frequently. C. Chronic leg ulcers—seen more frequently in older patients. D. Arthralgia—often with fever. E. Central nervous system disorders which may result from cerebral thrombosis. F. Cardiomegaly, both to right and left. G. Splenomegalia, usually in younger patients.

These patients frequently show an X-ray picture which is due to bone changes associated with chronic overactivity of the bone marrow. Many of these patients have an elongated skull. There may be a widening of the diploic spaces with prominent trabeculations giving a "sun-ray" appearance.

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II. Laboratory finds are: A. Red blood cell counts of 1,000,000 to 2,000,000. B. Hemoglobin reduced in accordance with the red blood cell volume. C. Sickling of the reduced, unstained blood. D. Reticulocytosis from 10.0% to 40.0%. E. Leukocytosis with a shift to the left and counts as high as 25,000. F. Platelet counts may be increased. G. Serum bilirubin usually elevated. H. Elevated icteric index. I. Urobilinogen elevated in feces and urine.

III. Treatment is symptomatic. Splenectomy is of little or no value with operative risk. Transfusions may be necessary during severe anemia and crisis.

IV. Prognosis: Patients with this condition have a poor prognosis with decreased life expectancy the rule. Death is due to infection or thrombosis of a vessel supplying a vital area.

V. Dental considerations: Patients with sickle cell anemia, by the very nature of their condition, create dental problems of which both the physician and the dentist should be aware. Medico-dental liaison is called for in these cases as it is in hemophiliacs, cardiacs, diabetics, etc.

Because of the greater susceptability to disease and infection and because of the greater surgical risk, these patients should be vigorously encouraged to have their teeth restored and saved.[3] Thoma and Robinson state that "infections accentuate the hemolytic symptoms and may result in death."[4] Thus the avoidance of infection is very important.

In the event that removal of teeth is necessary, certain precautions should be exercised. The patient's red blood cell count should be evaluated to determine if blood transfusion is indicated. In periods of remission or relative quiescence, these patients usually do not need transfusion prior to or following tooth extraction. Dental extraction should be performed during this time if at all possible. Since dental extraction is almost always an elective procedure, one would normally attempt to give paliative treatment with analgesics and/or antibiotics to a patient in a sickle cell crisis in need of an emergency dental extraction. Unfortunately, some of these patients may go for prolonged periods in crisis or sub-acute crisis and in these cases transfusion prior to extraction will probably be indicated.

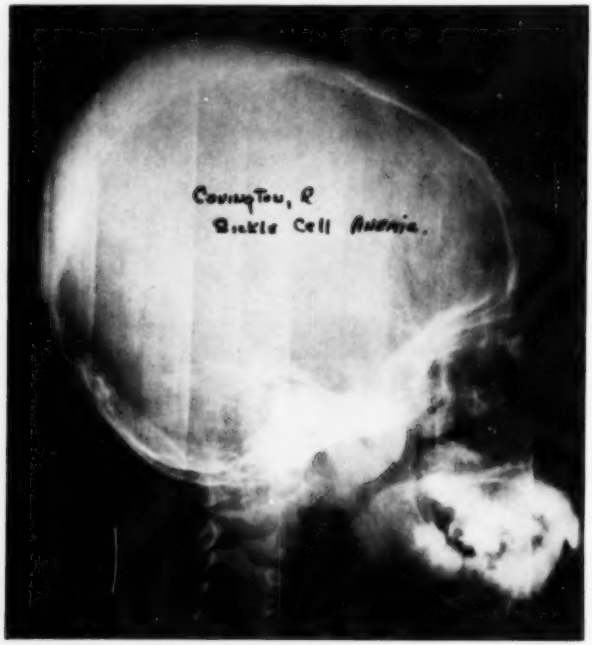

Fig. 1. Skull X-ray showing "hair-on-end" appearance, thin and porous anterior table and increased diploic spaces.

The choice of anesthesia is very important. Local infiltration or block anesthesia is preferred to general anesthesia. These patients have a decreased oxygen carrying capacity (which accounts for their general weakness, easy fatigability, and shortness of breath). Therefore, intravenous barbiturate anesthetics which depress central respiratory centers and inhalation agents which do not allow a high oxygen content are to be avoided. In addition, "drugs which are effective at high partial pressures, such as nitrous oxide, are hazardous when administered in the presence of anemia because of the possibility of asphyxia through technical error.[5] Also as "the maximum sickling occurs in areas in which there is anoxemia," it follows that sickle cell crisis may be precipitated by any form of even temporary anoxemia.[6] The general anesthetic of choice would be cyclopropane as it permits adequate oxygenation and is quickly liberated. Since this agent is usually unavailable in out-patient dental offices, if a general anesthetic is required, the patient should be hospitalized.

The decision as to the amount of dental surgery to be performed at any one appointment should be made by the dental surgeon. He should be acquainted with all the facts of the specific case as well as a thorough knowledge of the condition. This can best be accomplished by good medico-dental interchange of experiences and thought. "It is known that sickled cells are unusually sensitive to the effects of mechanical trauma in vitro and it is likely that this is an important mechanism causing destruction in vivo."[7] This being the case, surgical trauma should be minimized. Against this must be weighed the disadvantages of multiple procedures, multiple hospitalizations, multiple anesthetics, etc. Certainly a patient in a quiescent condition can withstand several extractions at one time while a patient in crisis might die as the result of a single extraction. Lastly, the decreased healing ability of the patients is still another reason for minimizing the number of areas exposed to surgery at any one time.

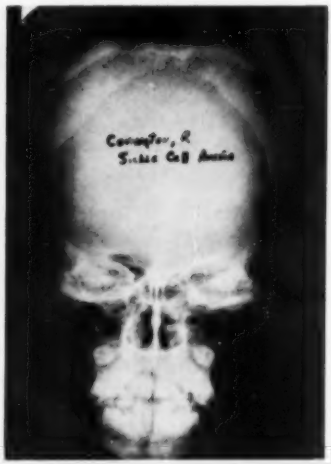

Fig. 2. X-ray of alveolor process and teeth of mandible showing increased radiolucency and "step-ladder" appearance of trabeculations.
Fig. 3. Posterior-anterior X-ray of skull showing greatly increased diploic spaces and "hair-on-end" appearance of trabeculations.

The dentist is in a position to suspect undiagnosed sickle cell anemia on two counts: a.) oral lesions and b.) radiographic findings. To this might be added a third criteria, physiognomy, which in these cases has been described as "a peculiar mongoloid facial appearance with high malar eminences and a short nose."[8]

The lesions in the mouth are non-specific and range from a pallor suggestive of anemias in general to erosions and ulcers. Like many other diseases with non-specific oral leisons, the findings are more indicative of something being wrong than tip-offs to a particular process.[9]

The X-ray picture is similar to that found in all congenital hemolytic anemias. The maxilla and the mandible show a thinning of the cortex and an increase of the medullary area. There is a widening of the trabeculations. This gives the picture of increased radiolucency. The maxillary sinuses may be smaller or even obliterated. The skull may show verticle trabeculations called "hair-on-end" appearance. The anterior table of the skull becomes thin and porous and the diploic spaces are increased. There is an increased radiolucency on the dental roentgenogram. "This change is noted especially in the alveolar process between the tooth roots where the trabeculation may appear as horizontal rows creating a step-ladder effect."[10]

SUMMARY

Patients with sickle cell anemia present certain problems in management of which the physican and the dentist must be aware. Judicious evaluation and careful surgery is required when dental extraction becomes necessary. These patients should be encouraged to restore their teeth and maintain good oral hygiene as surgery is to be avoided, if at all possible.

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Authors' Summaries...

MELNICK, J. L., BENYESH-MELNICK, M. AND BRENNAN, J. C.: Studies on Live Poliovirus Vaccine. Its Neurotropic Activity in monkeys and its Increased Neurovirulence after Multiplication in Vaccinated Children. (The Journal of the American Medical Association 171:1165, October 31, 1959).

Tests in monkeys of the live poliovirus vaccines currently being used in field trials have shown them to be more neurovirulent than was found in the laboratories of the proponents of the vaccines. The range of neurovirulence varied from the least virulent (Sabin's type *2*)—which reacted negatively after intracerebral injection but contained at least 2,500 monkey poliomyelitis doses per milliliter after intraspinal injection—to the most virulent (Lederle's types 1 and *2*) which contained at least 300 monkey doses per milliliter by intracerebral injection and 300,000 monkey doses per milliliter by intraspinal inoculation. The higher titers found by intraspinal injection seem to hinge on the use of a fine needle and the retention of the virus inoculum within the spinal cord.

The degree of genetic stability of the attenuated poliovirus as it undergoes multiplication cycles in vaccinated children and their contacts is of the utmost importance.[12] The use of tissue-culture markers for monitoring the stability of attenuated strains in the field is illustrated in a study of Sabin's strains fed to children in Mexico City. The excreted viruses which showed the greatest alteration in the tissue-culture *d* and *T* markers also showed the greatest increase in monkey neurovirulence (active after intracerebral injection, and only a few virus particles sufficing to paralyze monkeys after intraspinal injection). One altered strain was tested in a chimpanzee and found to be paralytogenic when inoculated intraspinally. That the increased monkey neurovirulence was not a laboratory artefact, brought on by cultivation of the virus under conditions favoring the growth of virulent particles, is shown by the finding that the few virus particles present in rectal swabs (10 to 60 tissue culture doses) were sufficient to paralyze monkeys inoculated intraspinally.

In the course of the Mexican study, it was found that many serologically susceptible children (i. e., free of poliomyelitis antibody) were resistant to infection by the vaccine. As such children were found to be already excreting a nonpoliomyelitis enterovirus at the time the live poliovirus vaccine was fed, the resistance has been attributed to the current infection with the nonpoliomyelitis enterovirus blocking the implantation of the vaccine virus. Interference of this sort might well limit the effectiveness of an orally given vaccine in areas where enterovirus infections are common.

The question of safety of the live poliovirus vaccine resolves itself on the frequency of disease in vaccinated children and their contacts, as compared with that anticipated from natural infection with wild polioviruses. In measuring the safety and effectiveness of live poliovirus vaccine, field investigators should consider not only the number of children fed but also the number already naturally immune and the number of poliomyelitis cases to be expected if the same population were exposed to virulent virus.

If caution was called for in 1954 and 1955 when the Salk vaccine was introduced into large-scale field use, then caution should also be the watchword now.

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